Juxta-adrenal Ancient Schwannoma: A Rare Retroperitoneal Tumor

Daniel A. Wollin, MD,[1] Ganesh Sivarajan, MD,[1] Pratibha Shukla, MD,[2] Jonathan Melamed, MD,[2] William C. Huang, MD,[1] Herbert Lepor, MD[1]

[1]Department of Urology, New York University School of Medicine, New York, NY; [2]Department of Pathology, New York University School of Medicine, New York, NY

Retroperitoneal schwannoma is a rare tumor that is often misdiagnosed as malignancy due to a concerning appearance on cross-sectional imaging. Pathology and immunohistochemistry form the gold standard for diagnosis; as such, local excision is the treatment of choice for this disease. We present two cases of juxta-adrenal ancient schwannoma that were treated with adrenalectomy and discuss the current literature regarding this entity.

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KEY WORDS

Ancient schwannoma • Adrenal tumor • Adrenalectomy

Schwannomas are tumors of the nerve sheath cells that surround peripheral nerves throughout the body. These tumors are typically benign in nature, although malignant transformation has been documented in rare cases. Schwannomas are most often found in women between the ages of 20 and 50 years; although they can be found throughout the peripheral nervous system, they seldom present in the retroperitoneum.[1] Less than 0.2% of incidental adrenal and periadrenal masses are eventually diagnosed as schwannoma,[2] and only 0.7% of schwannomas are found in the retroperitoneal space.[3,4] These tumors are classically well encapsulated, hypervascular, and can appear heterogeneous, making them difficult to distinguish from more concerning lesions through imaging studies alone.

Schwannomas, or neurilemmomas, originate from neural crest cells and are histologically

composed of spindle-shaped tumor cells organized in the Antoni A and Antoni B regions (cellularly dense and sparse, respectively).[5] The term *ancient schwannoma* was coined in 1951 by Ackerman and Taylor to describe an uncommon histologic subtype with only occasional tumor cells surrounded by a hyalinized matrix, more characteristic of the degeneration associated with a long-standing and slow-growing mass.[6]

Given the paucity of data regarding juxta-adrenal and other retroperitoneal ancient schwannomas, and their similar appearance to more invasive cancers, we discuss two cases of juxta-adrenal ancient schwannoma that initially presented as a malignant-appearing adrenal mass.

Case 1

A well-appearing 60-year-old woman was referred after abdominal ultrasound performed for elevated liver function test results demonstrated a 6-cm left adrenal mass. The patient was noted to be entirely asymptomatic from the mass, denying headaches, palpitations, weight loss, abdominal pain, or flank pain. The patient's past medical history was noncontributory with the exception of hypertension, which was well controlled on a single medication.

The patient's physical examination results were benign, without obvious palpable mass in the abdomen/flank or tenderness to palpation. Basic laboratory values were normal and functional workup of this adrenal mass showed no hormonal hypersecretion.

Further imaging studies were performed, including an abdominal computed tomography (CT) scan with contrast, which noted the left adrenal lesion as a 6.3-cm heterogeneous mass with hemorrhage and calcification most likely consistent with malignant neoplasm (Figure 1). Additionally, a 1.8-cm left para-aortic nodule was noted along the descending aorta in the thorax, consistent with metastatic disease versus neurogenic tumor (Figure 2). A high-resolution chest CT was then performed to further characterize the thoracic lesion, which noted the previously documented 1.8-cm nodule along with a 1-cm left paraspinal nodule at T7-8 (Figure 3).

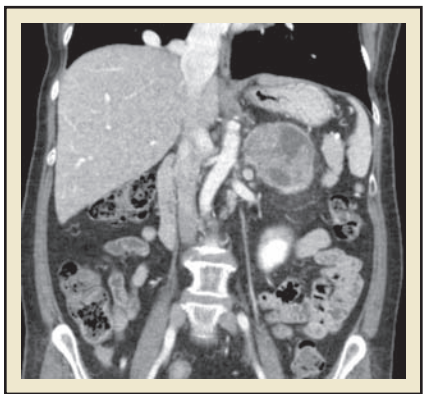

Figure 1. Computed tomography scan with contrast showing a 6.3-cm heterogeneous left adrenal lesion.

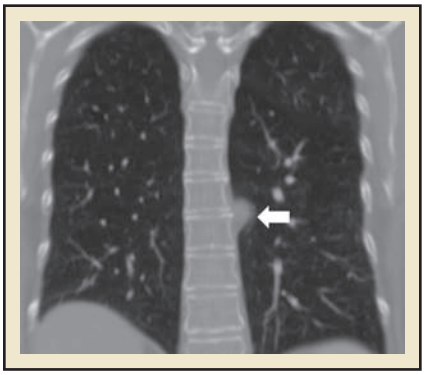

Figure 3. Noncontrast computed tomography scan of the thorax with the additional left paraspinal nodule (*arrow*).

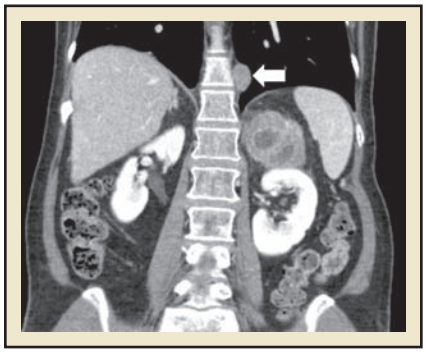

Figure 2. Computed tomography scan with contrast showing a left adrenal lesion along with left thoracic paraspinal nodule (*arrow*).

Given the appearance of the mass on CT scan and possible thoracic metastatic disease, the case was discussed at a multidisciplinary tumor board. The working diagnosis at this time was adrenocortical carcinoma and interventional radiology was consulted for biopsy of the thoracic lesions. Due to the unfavorable location, it was determined that the safest method for biopsy would be a thoracic surgery excisional biopsy, although this would be attempted after removal of the primary lesion. The consensus was that a cytoreductive adrenalectomy would improve the patient's symptom-free survival and possibly prolong her overall survival with adjuvant therapies, so the patient was scheduled for an open left adrenalectomy.[7]

The procedure was performed through a left anterior subcostal incision for optimal exposure of the tumor and surrounding tissues. Intraoperatively, the mass was noted to be densely adherent to the aorta, diaphragm, and left renal vasculature. Complete en bloc resection necessitated a left diaphragmatic entry (which was closed primarily) and a concomitant left nephrectomy. No additional gross disease was noted within the abdomen or retroperitoneum. After completion of the procedure, the patient recovered well and was discharged on postoperative day 4.

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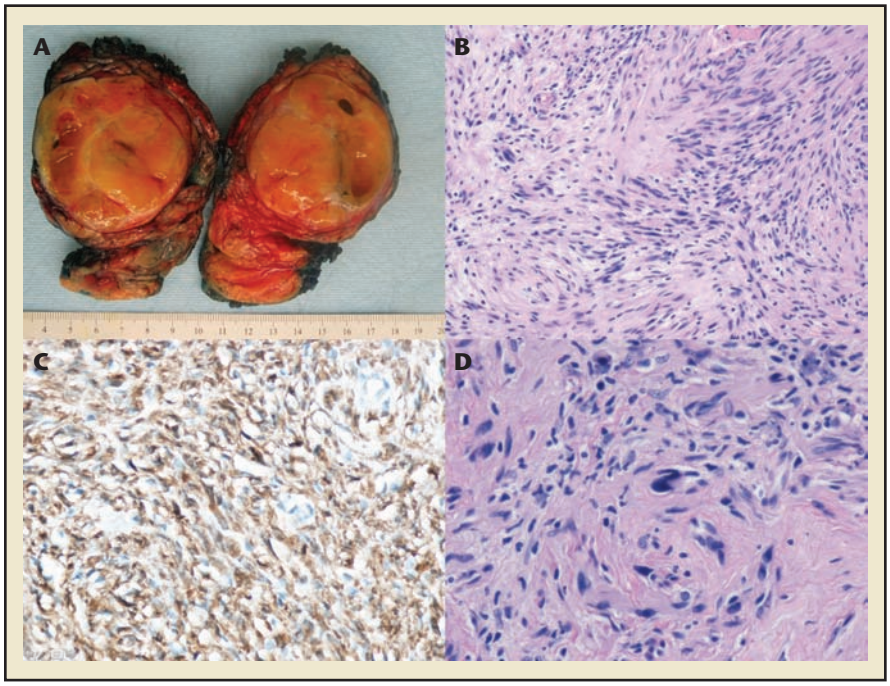

Figure 4. Juxta-adrenal schwannoma. **(A)** The tumor appears well encapsulated with adjacent adrenal tissue below, uninvolved with tumor. **(B)** Whorls of spindle cells arranged in fascicles with intervening stroma. **(C)** Schwannoma cells showing S100 immunoreactivity. **(D)** Scattered enlarged and hyperchromatic cells consistent with ancient change.

Pathologic examination of the resection specimen noted a 5.5-cm juxta-adrenal ancient schwannoma with an adjacent, and histologically normal, left adrenal gland. The mass itself was well encapsulated and yellow in appearance (Figure 4A). The tumor was composed of whorls of schwannoma spindle cells (Figure 4B), and had immunohistochemical activity for S100 protein, both consistent with schwannoma (Figure 4C). Additionally, the tumor showed significant signs of degeneration and contained scattered enlarged and hyperchromatic cells found in ancient schwannoma (Figure 4D). Additionally, six retroperitoneal lymph nodes and the left kidney were examined, all without signs of tumor invasion. After recovery, the patient was referred to thoracic surgery for management of the thoracic lesions, which were thought to be similarly neurogenic in origin with the current plan to reimage and determine subsequent need for resection.

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Case 2

A 43-year-old woman was discovered to have a large left adrenal mass during workup of nonspecific left flank pain for approximately 1 year. The patient had no prior medical or surgical history. An abdominal ultrasound was initially performed followed by a CT scan, both demonstrating a large (9.7 cm) left circumscribed mass of adrenal origin (Figure 5). Staging workup, which included a chest CT, showed no evidence of metastatic disease. Hormonal workup results were also negative, as in the previous case. Because of the large size of the mass and its adrenal origin, surgical excision was recommended, given the high likelihood of malignancy.

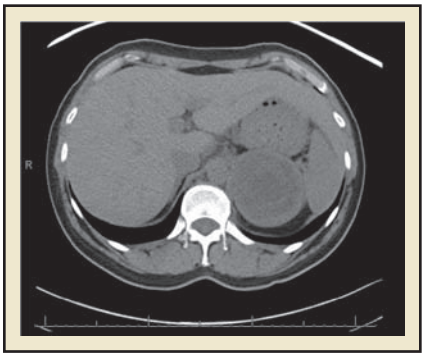

Figure 5. Computed tomography scan showing a 9.7-cm heterogeneous left adrenal lesion.

Robot-assisted laparoscopic left adrenalectomy was successfully performed via a transperitoneal approach. After ligation of the adrenal vein, the mass was successfully dissected free of the renal hilum and the upper pole of the kidney, such that concomitant nephrectomy was not necessary. The mass was, however, noted to be densely adherent to the aorta medially and the psoas muscle posteriorly. Several small vessels were noted to be feeding the large mass, each of which had to be individually clipped and cauterized. In order to ensure a negative margin, the mass was resected en bloc with a margin of the adherent, adjacent psoas fascia. The patient did well postoperatively and was discharged home on postoperative day 2.

Final pathology showed a 9.7 cm × 9.6 cm × 6.2 cm yellow-appearing mass with histologic characteristics of ancient schwannoma with extensive degenerative change. The cells were noted to have S100 staining, verified with immunohistochemical preparation, consistent with this diagnosis. Postoperatively she remains free of disease with a normal radiographic appearance of her left kidney at the time of her most recent follow-up.

Discussion

Schwannomas are tumors of the peripheral nerve sheath Schwann cells. As noted, this tumor is more common in women, with an approximate male-to-female ratio of 2:3.[3] They are rarely found in the retroperitoneum; less than 3% of benign schwannomas are seen in a retroperitoneal site.[8] Conversely, only approximately 6% of primary retroperitoneal tumors are found to be schwannomas.[9] Other than the increased risk of schwannomas associated with neurofibromatosis and similar genetic disorders, prior ionizing radiation is the only known modifiable risk factor for this disease, although increasing body mass index may possibly be protective.[10]

Given the slow-growing nature of many of these tumors, patients are often asymptomatic until the lesion becomes large enough to cause mass effect, stretching of tissues, and associated pain.[11] As patients undergo more frequent cross-sectional imaging and ultrasound, recent case reports have documented larger numbers of retroperitoneal schwannomas found incidentally, as with our first patient, rather than from the aforementioned symptoms, as seen with the second case. For this reason, they may be diagnosed at a smaller size.

Due to the frequent degeneration, cystic change, hemorrhage, and calcification within these masses, they often appear heterogeneous on cross-sectional imaging.[12] These tumors often have early weak enhancement with progressive heterogeneous and peripheral enhancement on delayed-phase CT and magnetic resonance imaging (MRI), both of which may lead to confusion with adrenal malignancy. On MRI, specifically, T1 hypointensity is common, along with T2 hyperintensity with a hypointense capsule.[12] The peripheral nerve site from which a schwannoma originates is not always visible on imaging, but the fibrous and occasionally enhancing capsule and calcifications are common signs of this tumor.[13,14]

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Due to the fact that ancient schwannomas cannot be easily diagnosed by imaging methods alone, the definitive diagnosis must be determined through histologic means.[6] The standard histologic characteristics of such tumors include areas of spindle-shaped tumor cells arranged in fascicles with minimal stromal matrix (Antoni A), along with areas of loosely packed myxoid lesions with occasional tumor cells, thought to be degeneration of the former areas (Antoni B). The collections of stromal matrix found within Antoni A areas between fascicles are known as *Verocay bodies* and are a typical histologic finding as well.[5] Immunohistochemical stains for the neuroectodermal marker S100 are nearly universally positive in these tumors.[15] Although nuclear palisades are a feature of classic schwannoma cells, these are typically absent in ancient schwannomas, which instead have nuclear atypia and hyperchromasia.[16]

Schwannomas are typically benign tumors, although malignant schwannomas do exist, most commonly in patients with neurofibromatosis.[17,18] Due to their overwhelmingly benign nature, treatment of choice for these masses is complete local tumor excision only; some advocate wide resection margins and sacrifice of nearby tissue if necessary as malignancy cannot be ruled out completely preoperatively.[19]

Although these benign tumors generally do not typcially invade adjacent organs, significant displacement of nearby structures frequently occurs. Retroperitoneal schwannomas can present in close proximity to the great vessels and with several feeding vessels draining from the tumor into the vena cava.[20] There have been reports of tumors displacing the rectosigmoid colon, ureter, uterus, and psoas muscle, as well as reports of a schwannoma eroding through a lumbar vertebra and encasing lumbar nerve roots.[21,22]

Although successful laparoscopic resection of retroperitoneal tumors near the great vessels has been described,[23,24] there may be an increased risk of hemorrhage, which can result in unsuccessful resection and even intraoperative mortality.[21] Moreover, it has been suggested that the laparoscopic resection of larger tumors ($>$ 5 cm) may result in hospital stays that are actually longer than with open resection.[19,20]

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Controversy exists over the utility of less aggressive surgical options for this benign lesion, such as simple enucleation, as recurrence rates with this method range from nonexistent to greater than 50%.[25,26] Considering the frequent concern for malignancy with ancient schwannoma due to the heterogeneous appearance on

imaging, radical excision is usually the recommended treatment in these situations. In our cases, malignancy was a strong consideration, and thus, en bloc resection of the mass along with surrounding tissue was required.

Conclusions

Retroperitoneal schwannoma is a rare entity, with a concerning appearance on cross-sectional imaging leading to frequent misdiagnosis. The "ancient" histologic subtype may occur in slow-growing and long-standing tumors with a diffuse hypocellularity within the specimen. The tumors are typically well encapsulated, although they can be densely adherent to surrounding structures, making complete resection (which is the treatment of choice) occasionally very challenging. This rare retroperitoneal entity should be considered in certain cases, with histologic and immunohistochemical evaluation as the gold standard for diagnosis. ■

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MAIN POINTS

- Schwannomas are tumors of the nerve sheath cells that surround peripheral nerves throughout the body. These tumors are typically benign in nature, although malignant transformation has been documented in rare cases. Although they can be found throughout the peripheral nervous system, they seldom present in the retroperitoneum.
- Given the slow-growing nature of many of these tumors, patients are often asymptomatic until the lesion becomes large enough to cause mass effect, stretching of tissues, and associated pain.
- Due to the fact that ancient schwannomas cannot be easily diagnosed by imaging methods alone, the definitive diagnosis must be determined through histologic means.
- Due to their overwhelmingly benign nature, treatment of choice for these masses is complete local tumor excision only; some advocate wide resection margins and sacrifice of nearby tissue if necessary as malignancy cannot be ruled out completely preoperatively. Although these benign tumors generally do not typically invade adjacent organs, significant displacement of nearby structures frequently occurs.